

Transcript

Does CWD pose a threat to human health?

James Kazmierczak, Epidemiologist, Wisconsin Department of Health

Hi, I'm Jim Kazmierczak. I'm with the state division of public health.

The single issue that I am going to try to address today is the issue of whether chronic wasting disease poses any risk to human health. As you probably already heard earlier in the presentation, chronic wasting disease belongs to a family of related diseases called *transmissible spongiform encephalopathies*, or TSE's for short. These diseases are related in the sense that they're all caused by this infectious protein that we call a prion, and also they're related in the fact affected the all present the same sort of clinical disease as a result of the type of lesions that they produce in the brain.

We know that different diseases in this family called TSE's, affect different species. For instance, we have disease called *scrapie* that affects sheep and goats. We have a particular kind of prion that affects mink. We have the bovine spongiform encephalopathy that's been in the news a lot, the Mad Cow Disease that affects obviously bovines. We have chronic wasting disease that affects deer and elk. And actually humans have their own unique type of spongiform encephalopathy. That disease is called Creutzfeldt-Jakob Disease, or CJD. This is the disease of humans that is similar, it's in the same group or family, as the other TSE's that we just mentioned.

I want to talk a bit about Creutzfeldt-Jakob Disease in humans. I feel I need to cover this because first of all, it's logical to assume that any prion disease that may in fact cross the species barrier to humans, will result in a similar type of disease as Creutzfeldt-Jakob Disease. Also, you're going to need to know a little bit about Creutzfeldt-Jakob Disease and in order to understand some of the points I'll make when I talk about the potential risk of chronic wasting disease to humans.

Creutzfeldt-Jakob Disease is a nasty disease. It produces progressive dementia, confusion, lack of coordination, and eventually death. This is an invariably fatal disease, and it usually kills its victims within twelve months of onset. The incubation period of this disease-- that is, the time between when one becomes exposed to the infectious protein, and when one actually develops symptoms, can be very variable, and it can range from between 15 months to over 20 years. So it can have an extraordinarily long incubation period.

As far as how Creutzfeldt-Jakob Disease is diagnosed in humans, the first thing you need to know is that there is no test that can be applied to show that someone is in the incubation phase of CJD. It's not until clinical signs appear that the disease can start to be diagnosed, and at that point the diagnosis is based on clinical signs and symptoms, laboratory tests, electroencephalogram patterns, and the definitive diagnosis is then made by actually looking at sections of the brain microscopically either with a brain biopsy or with a post-mortem examination.

I'm going to add another wrinkle to this. We've been looking at CJD or Creutzfeldt-Jakob Disease until now as a single entity-- when in fact one could split CJD into two different forms. The first is what we call classic Creutzfeldt-Jakob Disease. This form of the disease has been known to occur for at least 70 years, and it was probably occurring long before that. So this is not a new disease. It affects primarily older people. The vast majority of people to do get this disease are over the age of 65, and it affects people worldwide at a rate of approximately one case per million people. And that rate holds true here in the United States, including in Wisconsin.

How does one acquire this classic form of Creutzfeldt-Jakob Disease? In many cases it's just not known with certainty; although there are some mechanisms that are well-known. The majority of

cases do appear to occur spontaneously for no apparent reason. There are, however, a percentage of cases that apparently are familial; in other words, there is a family history of this, so there is a genetic component to a certain percentage of these cases, and interviewing patient families we can find a family history of this. It is also possible to acquire CJD through certain medical procedures; procedures that involve the transfer of tissues or tissue extracts from an infected patient (although obviously it's unknown to the medical providers at the time that the person was infected). But it can be transmitted, for instance, to people who receive pituitary extract; or certain brain covering tissue that is used for transplants. The infectious agent can be transmitted from the sick individuals to the well individuals by these sorts of medical procedures. And finally there's a small number of people who actually have developed this from ingestion. You may think that's an odd way of acquiring it, but this was discovered upon study of a Creutzfeldt-Jakob-like disease among some New Guinea tribesmen back several decades ago. At that time they were still practicing some ritualistic cannibalistic practices, and that's how we know the disease can also be transmitted by ingestion.

So that's sort of a summary of the classic form of Creutzfeldt-Jakob Disease. Again this is the form of disease has been known to occur for decades, and has been occurring worldwide at a rate of approximately one per million people. Now, what's new on the scene is a form of the disease called *new variant* Creutzfeldt-Jakob Disease, or new variant CJD. This form of the disease was first seen in England only about six years ago, and it is still limited to the United Kingdom and some European countries. It has not been found in the United States. New variant CJD is quite similar to the classic form that we've just finished talking, about it does tend to occur in younger people-- people with an average age in their thirties, actually-- and it differs in some ways-- subtly, though-- from the classic CJD in clinical signs. For instance, a neurologist would normally be able to differentiate on a clinical basis and some laboratory work, whether one has the classic form, or one has this new variant form of CJD. There's also enough difference in the microscopic appearance of brain tissue, so the bottom line is that the two types of disease-- the classic and the new variant forms-- can be differentiated.

So why has this new variant of Creutzfeldt-Jakob Disease appeared on the scene, seemingly of nowhere? Well, most of you probably know that during the 1980s there was an epidemic of bovine spongiform encephalopathy (better known as Mad Cow Disease) that occurred in Great Britain. It certainly got tons of media coverage at the time. Hundreds of thousands of cattle at that time were infected, many of which eventually found their way into the human food chain. The best evidence suggests that the prion that causes the bovine encephalopathy somehow managed to jump the species barrier and affect humans. Apparently when this prion did manage to infect a different species (in other words did manage to get in to humans), it produced a different clinical picture than the classic form of CJD.

By now you're probably wondering, "So what?" Now that we know the background about this disease form in humans, let's go back to the original question about the potential for chronic wasting disease to affect human beings. First of all, as far as we know there's never been the case of new variant Creutzfeldt-Jakob Disease here in this country-- and people have looked. The Centers for Disease Control began looking for the new variant form of Creutzfeldt-Jakob Disease several years ago. What they're doing is looking at people with compatible symptoms to Creutzfeldt-Jakob Disease who are under the age of 55. If you recall, I said that this disease tends to occur mostly in younger people, so onset at a young age would be a tip-off that we may be seeing this new variant form of Creutzfeldt-Jakob Disease.

The other bit of information that is somewhat reassuring as far as looking at the human health potential of chronic wasting disease, is that the classic form of CJD which does occur in this country, does not occur at any increased rates in the states out west that have had chronic wasting disease in their deer and elk herd for decades. Those states are not seeing any increase in Creutzfeldt-Jakob Disease, compared to states that don't have this disease in their deer herds. Furthermore, both the World Health Organization and the Centers for Disease Control state that there's no scientific evidence to prove any sort of link between chronic wasting disease and any

human health affects. And finally, researchers have never been able to demonstrate infectivity of muscle tissue, whether that's from CWD infected deer or elk, or BSE infected cattle, they've never been able to show that muscle tissues actually is infective as far as transmitting this disease.

So those are all fairly reassuring points, but-- as we mentioned earlier it appears that at least once in the past, a prion disease has been able to jump the species barrier and affect humans. This is what happened in Great Britain with the Mad Cow problem. And in a study that was just recently published, mice that were injected with a strain of the prion protein that was actually adapted to grow in rodents, they found that when they infected these mice they were actually able to transfer the infection by way of their skeletal muscle. So at least under these laboratory conditions, skeletal muscle can be infectious in those circumstances. Whether that's truly relatable to what happens in the real world, frankly we just aren't sure. So can one say with certainty that chronic wasting disease will never cause human disease? Unfortunately, not. None of us has a crystal ball, and it's actually very difficult, as you might expect, to be able to prove a negative-- to prove that CWD will never affect humans. I do think that if there is any risk-- and I said if-- it's likely to be quite low, based on the fact that first of all chronic wasting disease has never been known to affect humans, and also based on what we know about the epidemic of BSE in England and the new variant form of CJD in humans there. Here we are about a decade and a half after the outbreak in cattle, and even today there's only approximately 120 cases in all of Europe of the new variant Creutzfeldt-Jakob Disease. So that's fairly reassuring. I do wish I could quantify this risk in some way and try to put it in some perspective-- like perhaps comparing it to, say, the risk of cigarette smoking or the risk of drinking and driving. But it's impossible to try to quantify an event that's never occurred-- and again, CWD has never been known to result in human illness. As far as anyone knows, this is strictly a disease of deer and elk.

Finally, the question is, "Is it safe to eat venison from deer that are harvested from this area?" Even though there is no evidence that CWD has ever caused human disease, because of the uncertainty about how it's transmitted, experts in the World Health Organization *do* advise that no part of any deer or elk that's known to be infected with CWD, be consumed by people. It also suggests that people avoid consuming certain tissues of *any* deer or elk-- and that would include tissues like brain, spinal cord, eyes, spleen, tonsils and lymph nodes-- because that's where the prion tends to congregate, in those tissues. So although the World Health Organization does state that there's never been any scientific evidence to demonstrate that CWD can cause illness in humans, they do recommend not eating any venison from known infected deer. So are they hedging their bets? Well, sure they are-- even though the scientific evidence so far does not indicate a risk, no one can give you that absolute guarantee of future safety. The best that I could do is to try to accurately convey to you what's known about the potential risk, and then let each person decide how comfortable he or she is with that small degree of uncertainty about the safety of venison. Unfortunately there just are no easy answers